Executive Summary

In 1995, 865 cases of birth defects were detected among live born infants and fetuses of 20 or more weeks gestation delivered to mothers residing in Texas Public Health Regions 6 and 11 (Houston/Galveston region and Lower Rio Grande Valley). In this first full year of the Registry, surveillance was limited to approximately 23 major categories of birth defects, comprising 30 to 40 percent of all known structural malformations. Down syndrome, oral clefts, and spina bifida were the most common birth defects, although some major structural malformations were not yet monitored in 1995.

Age Patterns: Both trisomy 21 (Down syndrome) and trisomy 18 (Edwards syndrome) demonstrated an age-specific rate pattern that was J-shaped, with the highest birth prevalence ("rates") observed among mothers 35 years of age and older. Gastroschisis, a malformation of the abdominal wall, exhibited highest rates among the youngest mothers and decreased with each older age group.

Racial/Ethnic Patterns: Anencephaly and spina bifida were lowest among African Americans; however, rates were similar in whites and Hispanics. Among the heart defects, relatively high rates of tetralogy of Fallot were observed among African Americans, although they exhibited relatively low rates of hypoplastic left heart. Cleft palate alone was more likely to occur among whites and Hispanics. Rates of trisomy 18 (Edwards syndrome) were highest among African Americans and whites.

Gender Patterns: With regard to sex, higher rates were documented among females for anencephaly, and to a lesser extent, spina bifida. Females experienced two-fold higher rates of trisomy 18 (Edwards syndrome). Males had higher rates of cleft lip with or without cleft palate. However, for cleft palate alone, rates were similar for males and females.

Comparison with Other Registries: We compared Texas results with two other "benchmark" surveillance systems for birth defects (California and metropolitan Atlanta). Texas and Atlanta had higher rates of neural tube defects (NTDs, including anencephaly and spina bifida) than California. For oral clefts and trisomy 21 (Down syndrome), Texas and California recorded similar rates, both of which were somewhat higher than those observed for Atlanta.

Examination of the occurrence patterns of birth defects by region, sex, maternal age, and maternal race/ethnicity, as well as comparisons of Texas prevalence data with other surveillance systems, provide clues or research hypotheses for further studies, such as those conducted by the Texas Birth Defects Research Center.

History and Program Description

In April 1991, three infants were delivered in a 36-hour period in one facility in Brownsville, Texas with anencephaly, a birth defect in which much of the brain is missing. Astute clinicians recognized that this was excessive for this time period and facility, and they alerted the Texas Department of Health (TDH) of this cluster (an observed or reported excess of a health condition). Over the next couple of years, the Department, in cooperation with local officials and providers, as well as the Centers for Disease Control and Prevention (CDC), conducted a thorough epidemiologic investigation of neural tube defects (anencephaly and spina bifida) in Cameron County (which includes Brownsville) and Hidalgo County, the two most southeastern Texas counties that border Mexico. Compared with the United States, high rates of neural tube defects were confirmed for the area, especially Cameron County in 1991. The investigation underscored a general lack of background data on birth defects in Texas. In response to this cluster and need for better data, and in recognition of the enormous resources routinely put forth by the Department in the investigation of birth defects clusters statewide, the Texas State Legislature passed the Texas Birth Defects Act in 1993.

Out of this statute, the Texas Birth Defects Monitoring Division was created. The mission of this new Division is to identify and describe the patterns of birth defects in Texas, and to collaborate with others in finding causes of birth defects, working towards prevention, and linking families with services. This includes creating and maintaining the Texas Birth Defects Registry, monitoring for the excess occurrence of birth defects, conducting cluster investigations, and referring identified children and their families for services. The Texas Birth Defects Research Center was established in late 1996 at TDH through funding from the Centers for Disease Control and Prevention. Through this center, researchers will utilize registry cases to conduct epidemiologic studies in Texas and collaborate with seven other centers nationally to find preventable causes of birth defects.

This report summarizes our initial findings from the first full pilot year of the Texas Birth Defects Registry.

Methods

Case Definition

To be included as a case in the Texas Birth Defects Registry, all of the following must be true:

- The mother's residence at the time of delivery must be in an area covered by the Registry. Areas covered in 1995 are shown on the map.

- The infant/fetus must have a condition covered by the Registry. For 1995, these conditions include selected structural birth defects and fetal alcohol syndrome.

- The defect must be diagnosed or its signs or symptoms must be recognized within the first year of life. An exception is fetal alcohol syndrome, which must be diagnosed or recognized within the first six years of life.

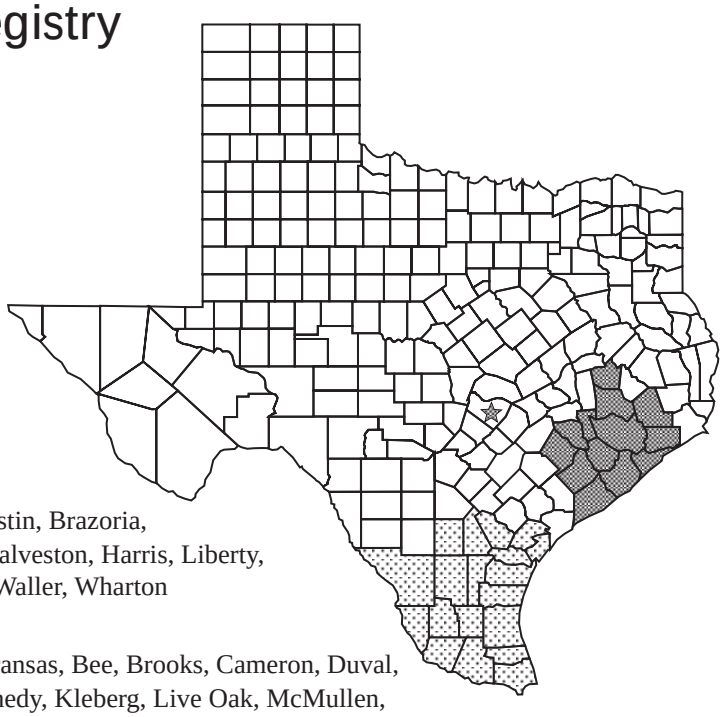
- The infant must have been born alive, or the fetus must have a gestational age of at least 20 weeks or a birth weight of at least 500 grams.

Pregnancies that end before 20 weeks are excluded from the case definition. Since some conditions may be prenatally diagnosed and the pregnancy terminated prior to 20 weeks, the observed rates may underestimate true occurrence. This is most likely to have an impact on anencephaly, spina bifida, trisomies 13, 18 and 21, and hydrocephalus.

The Registry also records information on additional events. Additional events are deliveries that do not meet the case definition, but are of interest. They include birth defects found among nonresident live births, and nonresident fetal deaths and pregnancy terminations of 20 or more weeks gestational age. ("Resident" means the mother was living in an area covered by the Registry at the time of delivery.) They also include birth defects found among resident pregnancy terminations prior to 20 weeks gestation.

To be included in this report, a case or additional event must have been delivered in 1995.

Texas Birth Defects Registry Pilot Project Areas



Public Health Region 6: Austin, Brazoria, Chambers, Colorado, Fort Bend, Galveston, Harris, Liberty, Matagorda, Montgomery, Walker, Waller, Wharton

Public Health Region 11: Aransas, Bee, Brooks, Cameron, Duval, Hidalgo, Jim Hogg, Jim Wells, Kenedy, Kleberg, Live Oak, McMullen, Nueces, Refugio, San Patricio, Starr, Webb, Willacy, Zapata

Data Collection Methods

The Texas Birth Defects Monitoring Division (TBDMD) uses active surveillance. This means it does not require reporting by hospitals or medical professionals. Instead, trained staff of the program routinely visit medical facilities where they have the authority to review logbooks, hospital discharge lists and other records. The staff member reviews the medical chart for each potential case identified. If the child has a birth defect covered by the Registry, detailed demographic and diagnostic information is abstracted. That information is entered into the computer and sent for processing. Quality control procedures for finding cases, abstracting information, and coding defects help ensure completeness and accuracy.

Data Analysis Methods

Results are presented for each type of defect covered in 1995, whether the defect occurred alone or together with others. Because a child often has more than one defect, it is not meaningful to sum over all diagnostic categories in the tables to obtain the total number of children with birth defects.

Tables include the number of cases found, the estimated prevalence per 10,000 live births, and the 95% confidence interval for the prevalence. Prevalence (sometimes referred to here as a rate) was calculated as follows:

$$\frac{\text{number of live births or fetal deaths with a birth defect}}{\text{total number of live births}} \times 10{,}000$$

The prevalence is only an estimate of the true prevalence, which is unknown. The confidence interval contains the true prevalence of a birth defect 95% of the time. A wide interval indicates the uncertainty stemming from small numbers. This report displays exact 95% confidence intervals based on the Poisson distribution. If one is comparing two prevalences and the 95% confidence intervals do not overlap, the prevalences are significantly different from each other. Furthermore, if two confidence intervals overlap substantially, the rates are probably not different statistically.

In this report, charts and text illustrate selected highlights of interest.

Birth Defects Data

Overall Prevalence at Birth

In 1995, there were 111,902 live births to residents of Public Health Regions 6 and 11, the pilot regions for the Texas Birth Defects Registry. A total of 865 cases were detected with one or more structural malformations in 1995. Of these, 776 were live born, corresponding to 0.7% of all live births. This is lower than the 3% rate commonly quoted for the U.S. (i.e., that 3% of all live births result in one or more major structural malformations). The difference is due primarily to the reduced case definition for 1995, which was limited to 23 selected major categories of malformations. The number of conditions monitored will expand in subsequent years.

In addition to live birth cases, 40 cases were detected among later fetal deaths (20+ weeks gestation) and 40 cases among induced pregnancy terminations that did not end in a live birth (also 20+ weeks). There were 9 other cases with other or unspecified pregnancy outcomes.

Trisomy 21, also known as Down syndrome, had the highest observed prevalence (12.96 cases per 10,000 live births) of any of the birth defects monitored by the Texas Birth Defects Registry in 1995. The prevalence of spina bifida, another well-known birth defect, was 5.36 per 10,000, with two-thirds of the cases having associated hydrocephalus. The prevalence for all other hydrocephalus was 4.74 per 10,000.

Only two definite cases of fetal alcohol syndrome (FAS) were detected during infancy. This is not an unusual finding, despite the relatively high prevalence of fetal alcohol exposure. It reflects the underdiagnosis and poor documentation of this condition during the neonatal and infancy periods.

Prevalence of Selected Birth Defects in the Area Covered by the Birth Defects Registry, Texas, 1995

Birth defect	Number of cases	Rate per 10,000 live births	95% confidence interval for rate
Central nervous system			
Anencephalus	32	2.86	1.96 - 4.04
Spina bifida with hydrocephalus	41	3.66	2.63 - 4.97
Spina bifida without hydrocephalus	19	1.70	1.02 - 2.65
Encephalocele	17	1.52	0.88 - 2.43
Microcephalus	45	4.02	2.93 - 5.38
Hydrocephalus	53	4.74	3.55 - 6.20
Cardiovascular and respiratory			
Transposition of the great vessels	51	4.56	3.39 - 5.99
Tetralogy of Fallot	26	2.32	1.52 - 3.40
Hypoplastic left heart	24	2.14	1.37 - 3.19
Agenesis or aplasia of lung	8	0.71	0.31 - 1.41
Oral clefts			
Cleft palate alone	74	6.61	5.19 - 8.30
Cleft lip with or without cleft palate	103	9.20	7.51 - 11.16
Gastrointestinal			
Tracheoesophageal fistula, esophageal atresia/stenosis	26	2.32	1.52 - 3.40
Atresia/stenosis of large intestine, rectum or anus	58	5.18	3.94 - 6.70
Genitourinary			
Renal agenesis or dysgenesis	56	5.00	3.78 - 6.50
Musculoskeletal			
Reduction defects of the upper limbs	24	2.14	1.37 - 3.19
Reduction defects of the lower limbs	14	1.25	0.68 - 2.10
Diaphragmatic hernia	24	2.14	1.37 - 3.19
Omphalocele	22	1.97	1.23 - 2.98
Gastroschisis	32	2.86	1.96 - 4.04
Chromosomal			
Trisomy 21 (Down syndrome)	145	12.96	10.93 - 15.25
Trisomy 13 (Patau syndrome)	13	1.16	0.62 - 1.99
Trisomy 18 (Edwards syndrome)	27	2.41	1.59 - 3.51
Other			
Fetal alcohol syndrome	2	0.18	0.02 - 0.65

Prevalence at Birth by Public Health Region

One should not place emphasis on interregional comparisons at this point, because our data collection methods were still being refined in 1995. Comparisons between regions will be more interesting in subsequent years, when data from several years can be combined to increase statistical power to detect differences.

Prevalence of Selected Birth Defects by Public Health Region, Texas, 1995

Birth defect	Public Health Region	Number of cases	Rate per 10,000 live births	95% confidence interval for rate
Central nervous system				
Anencephalus	6	24	3.18	2.04 - 4.74
	11	8	2.19	0.95 - 4.31
Spina bifida with hydrocephalus	6	20	2.65	1.62 - 4.10
	11	21	5.75	3.56 - 8.79
Spina bifida without hydrocephalus	6	12	1.59	0.82 - 2.78
	11	7	1.92	0.77 - 3.95
Encephalocele	6	9	1.19	0.55 - 2.27
	11	8	2.19	0.95 - 4.31
Microcephalus	6	32	4.25	2.90 - 5.99
	11	13	3.56	1.89 - 6.08
Hydrocephalus	6	38	5.04	3.57 - 6.92
	11	15	4.11	2.30 - 6.77
Cardiovascular and respiratory				
Transposition of the great vessels	6	28	3.72	2.47 - 5.37
	11	23	6.29	3.99 - 9.44
Tetralogy of Fallot	6	13	1.73	0.92 - 2.95
	11	13	3.56	1.89 - 6.08
Hypoplastic left heart	6	16	2.12	1.21 - 3.45
	11	8	2.19	0.95 - 4.31
Agenesis or aplasia of lung	6	8	1.06	0.46 - 2.09
	11	0	0.00	0.00 - 1.01
Oral clefts				
Cleft palate alone	6	46	6.10	4.47 - 8.14
	11	28	7.66	5.09 - 11.07
Cleft lip with or without cleft palate	6	72	9.55	7.48 - 12.03
	11	31	8.48	5.76 - 12.04
Gastrointestinal				
Tracheoesophageal fistula, esophageal atresia/stenosis	6	17	2.26	1.31 - 3.61
	11	9	2.46	1.13 - 4.68
Atresia/stenosis of large intestine, rectum or anus	6	35	4.64	3.23 - 6.46
	11	23	6.29	3.99 - 9.44

Prevalence of Selected Birth Defects by Public Health Region, Texas, 1995, continued

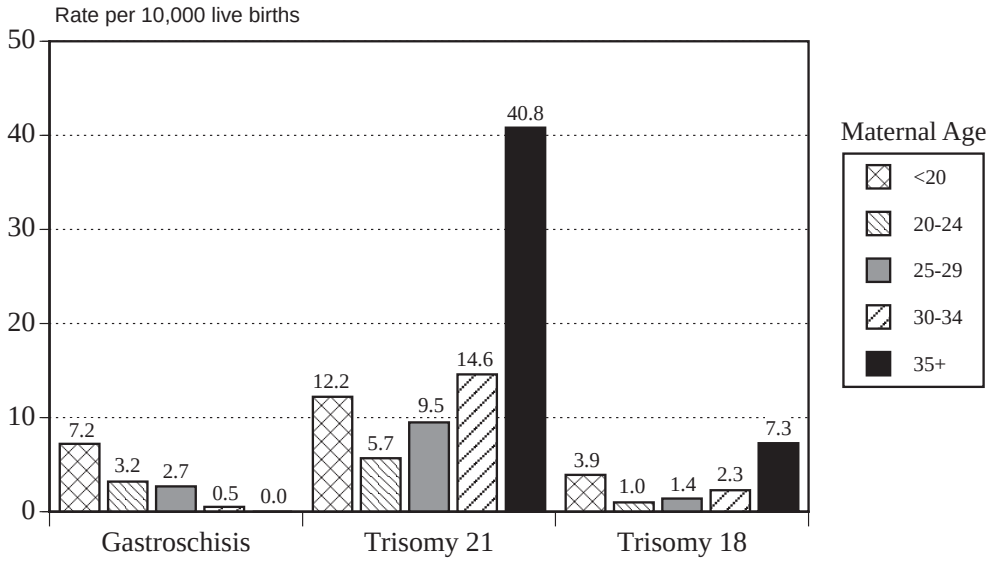
Birth defect	Public Health Region	Number of cases	Rate per 10,000 live births	95% confidence interval for rate
Genitourinary				
Renal agenesis or dysgenesis	6	36	4.78	3.35 - 6.61
	11	20	5.47	3.34 - 8.45
Musculoskeletal				
Reduction defects of the upper limbs	6	17	2.26	1.31 - 3.61
	11	7	1.92	0.77 - 3.95
Reduction defects of the lower limbs	6	13	1.73	0.92 - 2.95
	11	1	0.27	0.01 - 1.52
Diaphragmatic hernia	6	18	2.39	1.42 - 3.77
	11	6	1.64	0.60 - 3.57
Omphalocele	6	13	1.73	0.92 - 2.95
	11	9	2.46	1.13 - 4.68
Gastroschisis	6	21	2.79	1.72 - 4.26
	11	11	3.01	1.50 - 5.39
Chromosomal				
Trisomy 21 (Down syndrome)	6	100	13.27	10.80 - 16.14
	11	45	12.32	8.98 - 16.48
Trisomy 13 (Patau syndrome)	6	7	0.93	0.37 - 1.91
	11	6	1.64	0.60 - 3.57
Trisomy 18 (Edwards syndrome)	6	23	3.05	1.93 - 4.58
	11	4	1.09	0.30 - 2.80
Other				
Fetal alcohol syndrome	6	2	0.27	0.03 - 0.96
	11	0	0.00	0.00 - 1.01

Prevalence at Birth by Mother's Age

J-shaped age patterns were observed for trisomy 21 (Down syndrome) and trisomy 18 (Edwards syndrome). The oldest mothers were at greatest risk for delivering an infant or fetus with one of these chromosomal anomalies. Both conditions were roughly seven times more likely to occur in older mothers (35+ years of age) than in mothers 20-24 years old, the age group with the lowest rates. However, the trisomy 18 observations are based on a small number of cases.

For gastroschisis, a defect of the abdominal wall, mothers less than 20 years of age experienced the highest prevalence, with a decline in each subsequent age group. No age pattern was observed for omphalocele, another abdominal wall defect.

Prevalence of selected birth defects by mother's age at delivery among 1995 deliveries to residents of Texas Public Health Regions 6 & 11



Prevalence of Selected Birth Defects by Mother's Age, Texas 1995

Birth defect	Mother's age group	Number of cases	Rate per 10,000 live births	95% confidence interval for rate
Central nervous system				
Anencephalus	< 20	6	3.32	1.22 - 7.24
	20-24	11	3.50	1.75 - 6.26
	25-29	7	2.38	0.96 - 4.90
	30-34	6	2.74	1.00 - 5.95
	35+	2	1.81	0.22 - 6.55
Spina bifida with hydrocephalus	< 20	5	2.77	0.90 - 6.46
	20-24	13	4.14	2.20 - 7.07
	25-29	11	3.74	1.87 - 6.69
	30-34	9	4.10	1.88 - 7.79
	35+	3	2.72	0.56 - 7.94
Spina bifida without hydrocephalus	< 20	6	3.32	1.22 - 7.24
	20-24	4	1.27	0.35 - 3.26
	25-29	6	2.04	0.75 - 4.44
	30-34	2	0.91	0.11 - 3.29
	35+	1	0.91	0.02 - 5.05
Encephalocele	< 20	4	2.22	0.60 - 5.67
	20-24	4	1.27	0.35 - 3.26
	25-29	4	1.36	0.37 - 3.48
	30-34	3	1.37	0.28 - 4.00
	35+	2	1.81	0.22 - 6.55
Microcephalus	< 20	6	3.32	1.22 - 7.24
	20-24	15	4.77	2.67 - 7.87
	25-29	9	3.06	1.40 - 5.80
	30-34	8	3.65	1.57 - 7.19
	35+	7	6.34	2.55 - 13.07
Hydrocephalus	< 20	4	2.22	0.60 - 5.67
	20-24	14	4.45	2.43 - 7.47
	25-29	17	5.77	3.36 - 9.25
	30-34	12	5.47	2.83 - 9.56
	35+	6	5.44	2.00 - 11.83
Cardiovascular and respiratory				
Transposition of the great vessels	< 20	5	2.77	0.90 - 6.46
	20-24	13	4.14	2.20 - 7.07
	25-29	16	5.44	3.11 - 8.83
	30-34	11	5.01	2.50 - 8.97
	35+	6	5.44	2.00 - 11.83
Tetralogy of Fallot	< 20	5	2.77	0.90 - 6.46
	20-24	6	1.91	0.70 - 4.15
	25-29	9	3.06	1.40 - 5.80
	30-34	4	1.82	0.50 - 4.67
	35+	2	1.81	0.22 - 6.55

Birth defect	Mother's age group	Number of cases	Rate per 10,000 live births	95% confidence interval for rate
Hypoplastic left heart	< 20	6	3.32	1.22 - 7.24
	20-24	8	2.55	1.10 - 5.01
	25-29	5	1.70	0.55 - 3.96
	30-34	3	1.37	0.28 - 4.00
	35+	2	1.81	0.22 - 6.55
Agenesis or aplasia of lung	< 20	2	1.11	0.13 - 4.00
	20-24	0	0.00	0.00 - 1.17
	25-29	1	0.34	0.01 - 1.89
	30-34	2	0.91	0.11 - 3.29
	35+	3	2.72	0.56 - 7.94
Oral clefts				
Cleft palate alone	< 20	8	4.43	1.91 - 8.73
	20-24	20	6.36	3.89 - 9.83
	25-29	14	4.76	2.60 - 7.98
	30-34	23	10.49	6.65 - 15.73
	35+	9	8.16	3.73 - 15.48
Cleft lip with or without cleft palate	< 20	7	3.88	1.56 - 7.99
	20-24	37	11.77	8.29 - 16.22
	25-29	33	11.21	7.72 - 15.74
	30-34	16	7.29	4.17 - 11.85
	35+	10	9.06	4.35 - 16.66
Gastrointestinal				
Tracheoesophageal fistula, esophageal atresia/stenosis	< 20	4	2.22	0.60 - 5.67
	20-24	8	2.55	1.10 - 5.01
	25-29	8	2.72	1.17 - 5.35
	30-34	4	1.82	0.50 - 4.67
	35+	2	1.81	0.22 - 6.55
Atresia/stenosis of large intestine, rectum or anus	< 20	12	6.65	3.44 - 11.61
	20-24	16	5.09	2.91 - 8.27
	25-29	16	5.44	3.11 - 8.83
	30-34	10	4.56	2.19 - 8.38
	35+	4	3.62	0.99 - 9.28
Genitourinary				
Renal agenesis or dysgenesis	< 20	10	5.54	2.66 - 10.19
	20-24	17	5.41	3.15 - 8.66
	25-29	15	5.10	2.85 - 8.40
	30-34	10	4.56	2.19 - 8.38
	35+	4	3.62	0.99 - 9.28
Musculoskeletal				
Reduction defects of the upper limbs	< 20	4	2.22	0.60 - 5.67
	20-24	5	1.59	0.52 - 3.71
	25-29	5	1.70	0.55 - 3.96
	30-34	7	3.19	1.28 - 6.58
	35+	3	2.72	0.56 - 7.94

Prevalence of Selected Birth Defects by Mother's Age, Texas, 1995, continued

Birth defect	Mother's age group	Number of cases	Rate per 10,000 live births	95% confidence interval for rate
Reduction defects of the lower limbs	< 20	2	1.11	0.13 - 4.00
	20-24	5	1.59	0.52 - 3.71
	25-29	4	1.36	0.37 - 3.48
	30-34	1	0.46	0.01 - 2.54
	35+	2	1.81	0.22 - 6.55
Diaphragmatic hernia	< 20	6	3.32	1.22 - 7.24
	20-24	7	2.23	0.90 - 4.59
	25-29	4	1.36	0.37 - 3.48
	30-34	5	2.28	0.74 - 5.32
	35+	2	1.81	0.22 - 6.55
Omphalocele	< 20	2	1.11	0.13 - 4.00
	20-24	8	2.55	1.10 - 5.01
	25-29	5	1.70	0.55 - 3.96
	30-34	4	1.82	0.50 - 4.67
	35+	3	2.72	0.56 - 7.94
Gastroschisis	< 20	13	7.20	3.83 - 12.32
	20-24	10	3.18	1.53 - 5.85
	25-29	8	2.72	1.17 - 5.35
	30-34	1	0.46	0.01 - 2.54
	35+	0	0.00	0.00 - 3.34
Chromosomal				
Trisomy 21 (Down syndrome)	< 20	22	12.19	7.64 - 18.45
	20-24	18	5.73	3.39 - 9.05
	25-29	28	9.51	6.32 - 13.75
	30-34	32	14.59	9.98 - 20.59
	35+	45	40.78	29.74 - 54.56
Trisomy 13 (Patau syndrome)	< 20	0	0.00	0.00 - 2.04
	20-24	1	0.32	0.01 - 1.77
	25-29	5	1.70	0.55 - 3.96
	30-34	5	2.28	0.74 - 5.32
	35+	2	1.81	0.22 - 6.55
Trisomy 18 (Edwards syndrome)	< 20	7	3.88	1.56 - 7.99
	20-24	3	0.95	0.20 - 2.79
	25-29	4	1.36	0.37 - 3.48
	30-34	5	2.28	0.74 - 5.32
	35+	8	7.25	3.13 - 14.28
Other				
Fetal alcohol syndrome	< 20	0	0.00	0.00 - 2.04
	20-24	0	0.00	0.00 - 1.17
	25-29	0	0.00	0.00 - 1.25
	30-34	2	0.91	0.11 - 3.29
	35+	0	0.00	0.00 - 3.34

Prevalence at Birth by Mother's Race/Ethnicity

Race/ethnic groups are defined in this report using the approach of the Bureau of Vital Statistics, Texas Department of Health. The infant is assigned the group reported for the mother in the medical record. "White" is white, non-Hispanic. "African American" includes all African Americans, regardless of ethnicity. "Hispanic" includes people whose race is white, American Indian or other/non-classifiable, and have a Spanish-speaking country of origin. The "other" ethnic category in the table includes both mothers of unknown race/ethnicity and mothers of racial/ethnic groups other than white, African American, or Hispanic. Because of this mixture, one should not place much emphasis on the "other" category at this time.

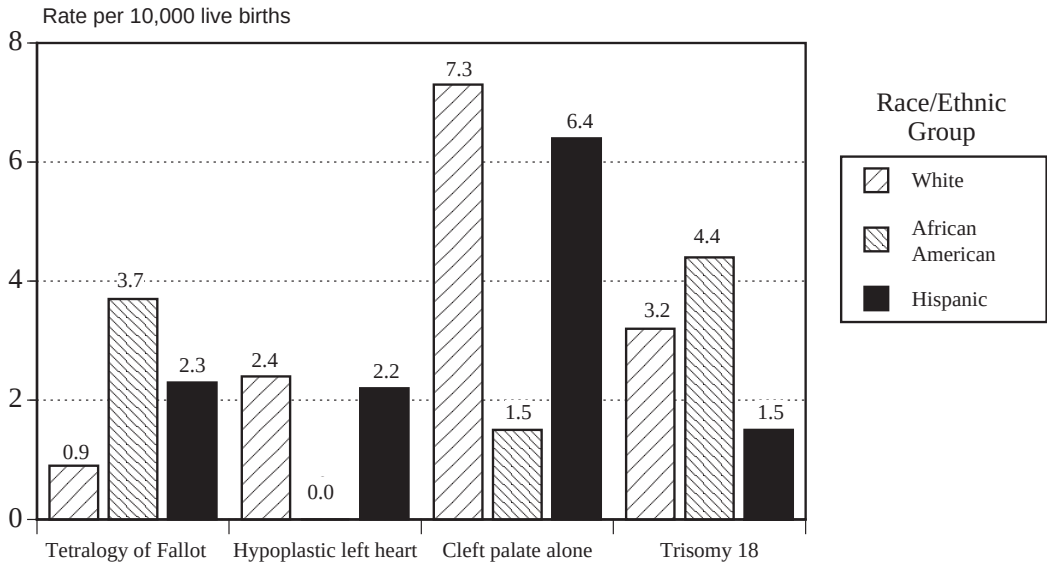
The birth prevalence of tetralogy of Fallot (a heart defect) was higher among African Americans than among whites. Higher rates of hypoplastic left heart were observed for whites and Hispanics than among African Americans. However, the total number of cases for these two heart defects was small.

Cleft palate alone (without cleft lip), was four to five times more likely to occur among whites and Hispanics than among African Americans, although there were only two cases in the latter group. African Americans also experienced lower rates of cleft lip with or without cleft palate, although the ethnic differences were not as pronounced as in the cleft palate category.

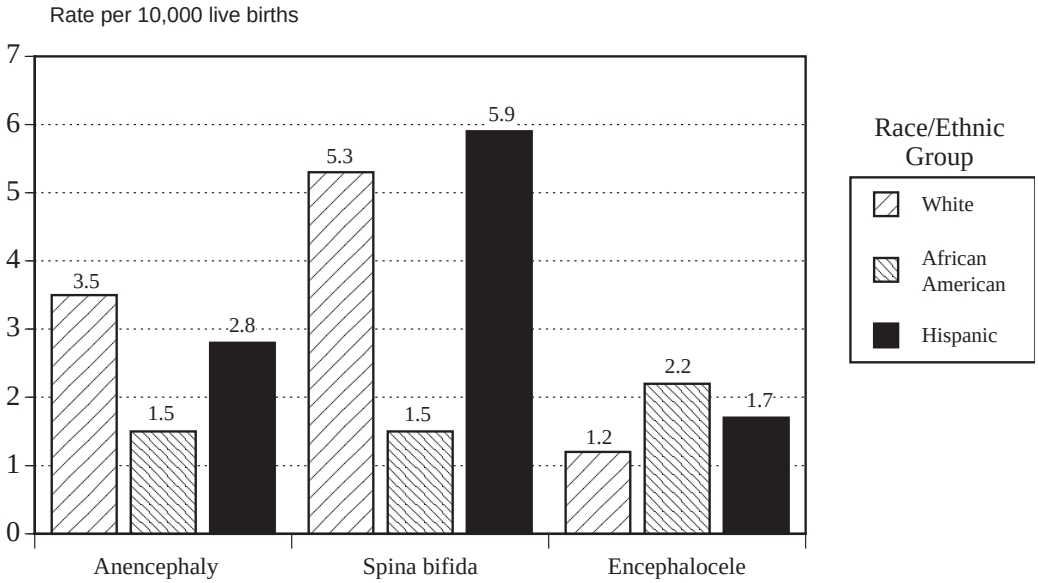
Rates for trisomy 18 (Edwards syndrome) were higher among African Americans and whites than among Hispanics. The reverse pattern was observed for trisomy 21 (Down syndrome), for which Hispanics had the highest rates.

Ethnic-specific rates are illustrated for neural tube defects (NTDs), which include anencephaly, spina bifida, and encephalocele. Spina bifida was the most prevalent NTD and encephalocele was the least prevalent among Hispanics and whites. African Americans experienced relatively low rates for anencephaly and spina bifida. Numerous studies have documented low NTD rates in African Americans, and high rates in Hispanics, relative to whites. However, no excess of cases among Hispanics was observed in our 1995 data.

Prevalence of selected birth defects by mother's race/ethnic group among 1995 deliveries to residents of Texas Public Health Regions 6 & 11



Prevalence of neural tube defects by mother's race/ethnic group among 1995 deliveries to residents of Texas Public Health Regions 6 & 11



Prevalence of Selected Birth Defects by Mother's Race/Ethnicity, Texas, 1995

Birth defect	Mother's race/ethnicity	Number of cases	Rate per 10,000 live births	95% confidence interval for rate
Central nervous system				
Anencephalus	White	12	3.52	1.82 - 6.15
	African American	2	1.46	0.18 - 5.28
	Hispanic	17	2.81	1.64 - 4.49
	Other	1	2.80	0.07 - 15.62
Spina bifida with hydrocephalus	White	11	3.23	1.61 - 5.77
	African American	2	1.46	0.18 - 5.28
	Hispanic	26	4.29	2.80 - 6.29
	Other	2	5.61	0.68 - 20.25
Spina bifida without hydrocephalus	White	7	2.05	0.83 - 4.23
	African American	0	0.00	0.00 - 2.70
	Hispanic	10	1.65	0.79 - 3.04
	Other	2	5.61	0.68 - 20.25
Encephalocele	White	4	1.17	0.32 - 3.00
	African American	3	2.19	0.45 - 6.41
	Hispanic	10	1.65	0.79 - 3.04
	Other	0	0.00	0.00 - 10.34
Microcephalus	White	16	4.69	2.68 - 7.62
	African American	10	7.31	3.50 - 13.44
	Hispanic	17	2.81	1.64 - 4.49
	Other	2	5.61	0.68 - 20.25
Hydrocephalus	White	24	7.04	4.51 - 10.48
	African American	8	5.85	2.52 - 11.52
	Hispanic	20	3.30	2.02 - 5.10
	Other	1	2.80	0.07 - 15.62
Cardiovascular and respiratory				
Transposition of the great vessels	White	20	5.87	3.58 - 9.06
	African American	4	2.92	0.80 - 7.49
	Hispanic	25	4.13	2.67 - 6.09
	Other	2	5.61	0.68 - 20.25
Tetralogy of Fallot	White	3	0.88	0.18 - 2.57
	African American	5	3.65	1.19 - 8.53
	Hispanic	14	2.31	1.26 - 3.88
	Other	4	11.21	3.06 - 28.71
Hypoplastic left heart	White	8	2.35	1.01 - 4.62
	African American	0	0.00	0.00 - 2.70
	Hispanic	13	2.15	1.14 - 3.67
	Other	3	8.41	1.73 - 24.58

Prevalence of Selected Birth Defects by Mother's Race/Ethnicity, Texas, 1995, continued

Birth defect	Mother's race/ethnicity	Number of cases	Rate per 10,000 live births	95% confidence interval for rate
Agenesis or aplasia of lung	White	4	1.17	0.32 - 3.00
	African American	1	0.73	0.02 - 4.07
	Hispanic	3	0.50	0.10 - 1.45
	Other	0	0.00	0.00 - 10.34
Oral clefts				
Cleft palate alone	White	25	7.33	4.75 - 10.83
	African American	2	1.46	0.18 - 5.28
	Hispanic	39	6.44	4.58 - 8.80
	Other	8	22.43	9.68 - 44.19
Cleft lip with or without cleft palate	White	34	9.97	6.91 - 13.94
	African American	8	5.85	2.52 - 11.52
	Hispanic	52	8.59	6.41 - 11.26
	Other	9	25.23	11.54 - 47.90
Gastrointestinal				
Tracheoesophageal fistula, esophageal atresia/stenosis	White	11	3.23	1.61 - 5.77
	African American	1	0.73	0.02 - 4.07
	Hispanic	13	2.15	1.14 - 3.67
	Other	1	2.80	0.07 - 15.62
Atresia/stenosis of large intestine, rectum or anus	White	11	3.23	1.61 - 5.77
	African American	2	1.46	0.18 - 5.28
	Hispanic	35	5.78	4.03 - 8.04
	Other	10	28.03	13.44 - 51.56
Genitourinary				
Renal agenesis or dysgenesis	White	22	6.45	4.04 - 9.77
	African American	6	4.39	1.61 - 9.55
	Hispanic	25	4.13	2.67 - 6.09
	Other	3	8.41	1.73 - 24.58
Musculoskeletal				
Reduction defects of the upper limbs	White	9	2.64	1.21 - 5.01
	African American	3	2.19	0.45 - 6.41
	Hispanic	12	1.98	1.02 - 3.46
	Other	0	0.00	0.00 - 10.34
Reduction defects of the lower limbs	White	4	1.17	0.32 - 3.00
	African American	3	2.19	0.45 - 6.41
	Hispanic	6	0.99	0.36 - 2.16
	Other	1	2.80	0.07 - 15.62
Diaphragmatic hernia	White	6	1.76	0.65 - 3.83
	African American	2	1.46	0.18 - 5.28
	Hispanic	15	2.48	1.39 - 4.08
	Other	1	2.80	0.07 - 15.62

Birth defect	Mother's race/ethnicity	Number of cases	Rate per 10,000 live births	95% confidence interval for rate
Omphalocele	White	5	1.47	0.48 - 3.42
	African American	4	2.92	0.80 - 7.49
	Hispanic	10	1.65	0.79 - 3.04
	Other	3	8.41	1.73 - 24.58
Gastroschisis	White	11	3.23	1.61 - 5.77
	African American	2	1.46	0.18 - 5.28
	Hispanic	18	2.97	1.76 - 4.70
	Other	1	2.80	0.07 - 15.62
Chromosomal				
Trisomy 21 (Down syndrome)	White	39	11.44	8.14 - 15.64
	African American	13	9.50	5.06 - 16.25
	Hispanic	88	14.53	11.65 - 17.90
	Other	5	14.02	4.55 - 32.71
Trisomy 13 (Patau syndrome)	White	4	1.17	0.32 - 3.00
	African American	1	0.73	0.02 - 4.07
	Hispanic	7	1.16	0.46 - 2.38
	Other	1	2.80	0.07 - 15.62
Trisomy 18 (Edwards syndrome)	White	11	3.23	1.61 - 5.77
	African American	6	4.39	1.61 - 9.55
	Hispanic	9	1.49	0.68 - 2.82
	Other	1	2.80	0.07 - 15.62
Other				
Fetal alcohol syndrome	White	0	0.00	0.00 - 1.08
	African American	2	1.46	0.18 - 5.28
	Hispanic	0	0.00	0.00 - 0.61
	Other	0	0.00	0.00 - 10.34

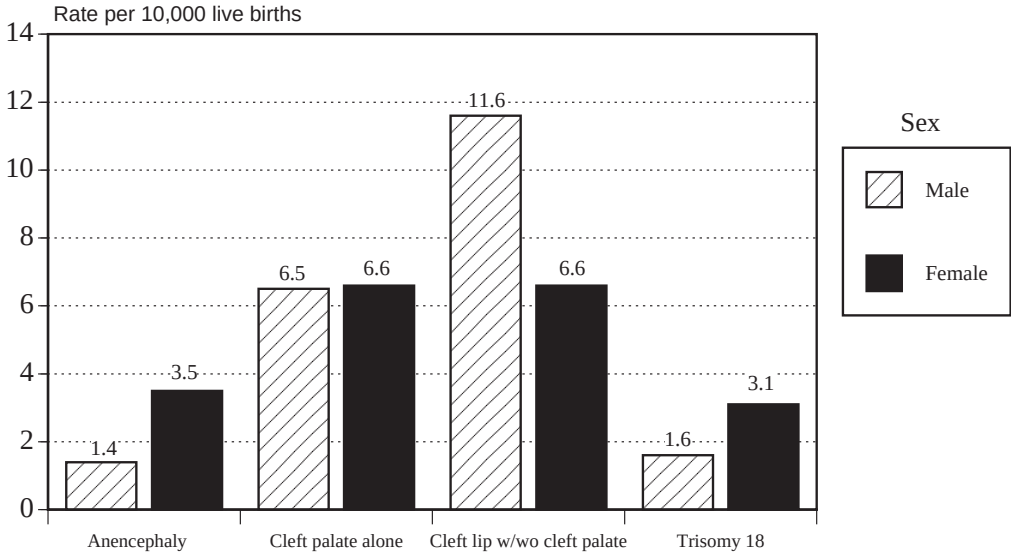
Prevalence at Birth by Sex

The rate of anencephaly was 2 1/2 times higher among female infants and fetuses than among males. The higher proportion of female anencephaly cases has been consistently documented in the literature and in other surveillance systems. Females had 54% higher rates for spina bifida (with and without hydrocephalus combined). Three spina bifida cases were of indeterminate sex. The rate of encephalocele, the other listed neural tube defect, was 49% higher among females than males.

For oral clefts, different sex patterns were seen, depending on the cleft category. Males and females had similar rates of cleft palate alone, but males were 76% more likely to have cleft lip with or without cleft palate.

The rate of trisomy 18 (Edwards syndrome) among females was nearly twice the rate among males. In contrast, females and males experienced similar rates for trisomy 21 (Down syndrome).

Prevalence of selected birth defects by sex among 1995 deliveries to residents of Texas Public Health Regions 6 & 11



Prevalence of Selected Birth Defects by Sex
Texas, 1995

Birth defect	Sex	Number of cases	Rate per 10,000 live births	95% confidence interval for rate
Central nervous system				
Anencephalus	Male	8	1.40	0.60 - 2.76
	Female	19	3.47	2.09 - 5.42
Spina bifida with hydrocephalus	Male	18	3.15	1.87 - 4.98
	Female	21	3.83	2.37 - 5.86
Spina bifida without hydrocephalus	Male	5	0.88	0.28 - 2.04
	Female	13	2.37	1.26 - 4.06
Encephalocele	Male	7	1.23	0.49 - 2.53
	Female	10	1.83	0.88 - 3.36
Microcephalus	Male	22	3.85	2.41 - 5.83
	Female	23	4.20	2.66 - 6.30
Hydrocephalus	Male	31	5.43	3.69 - 7.70
	Female	21	3.83	2.37 - 5.86
Cardiovascular and respiratory				
Transposition of the great vessels	Male	29	5.08	3.40 - 7.29
	Female	22	4.02	2.52 - 6.08
Tetralogy of Fallot	Male	13	2.28	1.21 - 3.89
	Female	13	2.37	1.26 - 4.06
Hypoplastic left heart	Male	13	2.28	1.21 - 3.89
	Female	11	2.01	1.00 - 3.59
Agenesis or aplasia of lung	Male	5	0.88	0.28 - 2.04
	Female	3	0.55	0.11 - 1.60
Oral clefts				
Cleft palate alone	Male	37	6.48	4.56 - 8.93
	Female	36	6.57	4.60 - 9.10
Cleft lip with or without cleft palate	Male	66	11.56	8.94 - 14.70
	Female	36	6.57	4.60 - 9.10
Gastrointestinal				
Tracheoesophageal fistula, esophageal atresia/stenosis	Male	13	2.28	1.21 - 3.89
	Female	13	2.37	1.26 - 4.06
Atresia/stenosis of large intestine, rectum or anus	Male	27	4.73	3.12 - 6.88
	Female	29	5.29	3.54 - 7.60

Prevalence of Selected BIrth Defects by Sex, Texas, 1995, continued

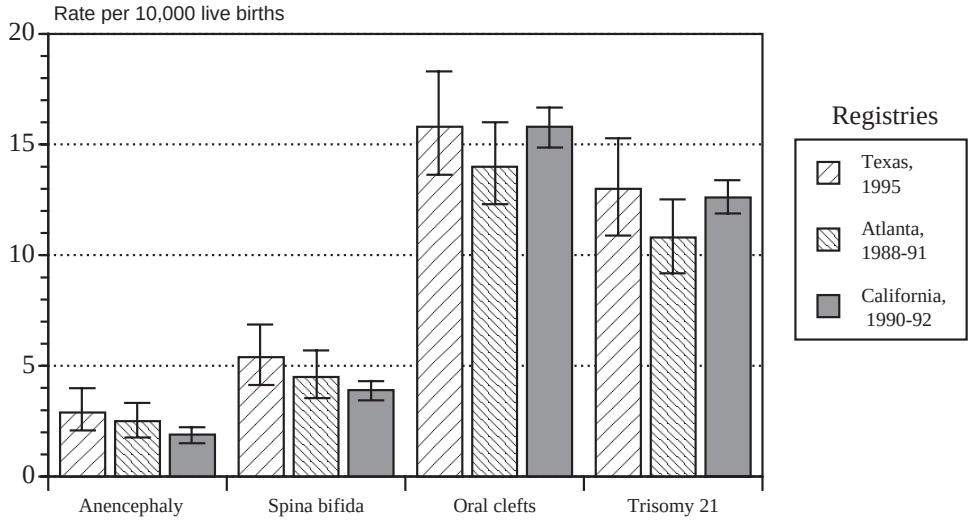
Birth defect	Sex	Number of cases	Rate per 10,000 live births	95% confidence interval for rate
Genitourinary				
Renal agenesis or dysgenesis	Male	29	5.08	3.40 - 7.29
	Female	24	4.38	2.81 - 6.52
Musculoskeletal				
Reduction defects of the upper limbs	Male	15	2.63	1.47 - 4.33
	Female	9	1.64	0.75 - 3.12
Reduction defects of the lower limbs	Male	3	0.53	0.11 - 1.54
	Female	9	1.64	0.75 - 3.12
Diaphragmatic hernia	Male	13	2.28	1.21 - 3.89
	Female	11	2.01	1.00 - 3.59
Omphalocele	Male	12	2.10	1.09 - 3.67
	Female	9	1.64	0.75 - 3.12
Gastroschisis	Male	15	2.63	1.47 - 4.33
	Female	16	2.92	1.67 - 4.74
Chromosomal				
Trisomy 21 (Down syndrome)	Male	71	12.43	9.71 - 15.68
	Female	73	13.32	10.44 - 16.75
Trisomy 13 (Patau syndrome)	Male	5	0.88	0.28 - 2.04
	Female	8	1.46	0.63 - 2.88
Trisomy 18 (Edwards syndrome)	Male	9	1.58	0.72 - 2.99
	Female	17	3.10	1.81 - 4.97
Other				
Fetal alcohol syndrome	Male	1	0.18	0.00 - 0.98
	Female	1	0.18	0.00 - 1.02

NOTE: the sum of birth defects among males and females may not equal the sum of birth defects shown in other tables, due to deliveries of undetermined sex.

Texas Data Compared with Data from Other Surveillance Systems

Texas Registry data were compared with data from two other benchmark surveillance systems for birth defects. With respect to neural tube defects, Texas (1995) and Atlanta (1988-91) had higher rates than California (1990-92) for both anencephaly and spina bifida. This might be partially explained by the fact that the California data represent cases found only among live births, but not among fetal deaths and induced terminations. There are also other slight differences in case definition. For oral clefts and trisomy 21 (Down syndrome), Texas and California had similar rates, both of which were higher than those observed for Atlanta. These are only crude comparisons, and do not adjust for differing maternal age distributions or differences in case definition.

Prevalence of selected birth defects in Texas and other registries



Texas and Atlanta: birth defects among live births and fetuses 20+weeks; California: birth defects among live births only.

Birth Defect Cluster Investigations in 1995

There were ten cluster investigations conducted in 1995*.

Area	Condition of of concern	Background	Response

*A more detailed report is available from the Texas Birth Defects Monitoring Division, at (512) 458-7232.

Appendix A:

Glossary

Agenesis Defective development or absence of part(s) of the body.

Agenesis, hypoplasia, and dysplasia of the lung The absence or incomplete development of a lung.

Anencephalus Congenital absence of the skull, with cerebral hemispheres completely missing or reduced to small masses attached to the base of the skull.

Atresia Imperforation; absence or closure of a normal opening or hollow organ.

Atresia and stenosis of large intestine, rectum and anal canal The absence, closure or constriction of the large intestine, rectum or anal canal.

Cleft palate The congenital failure of the palate to fuse properly, forming a grooved depression or fissure in the roof of the mouth.

Cleft lip The congenital failure of the maxillary and median nasal processes to fuse, forming a groove or fissure in the lip.

Confidence interval (95%) The interval that contains the true prevalence (which we can only estimate) 95% of the time. See Methods for more explanation.

Congenital Existing at or dating from birth.

Diaphragmatic hernia The protrusion of an abdominal organ through a defect in the diaphragm.

Down syndrome See Trisomy 21.

Edwards syndrome See Trisomy 18.

Encephalocele The protrusion of the brain substance through a defect in the skull.

Fetal alcohol syndrome A constellation of physical abnormalities (including characteristic abnormal facial features and growth retardation), and problems of behavior and cognition in children born to mothers who drank alcohol during pregnancy.

Gastroschisis A congenital opening of the abdominal wall with protrusion of the intestines.

Hydrocephalus The abnormal accumulation of fluid within the skull.

Hyperplasia An abnormal or unusual increase in the elements composing a part (as tissue cells).

Hypoplasia A condition of arrested development in which an organ or part remains below the normal size or in an immature state.

Hypoplastic left heart syndrome Atresia, or marked hypoplasia, of the aortic opening or valve, with hypoplasia of the ascending aorta and defective development of the left ventricle (with mitral valve atresia).

Limb defects See Reduction deformities.

Meninges Membranes that cover the brain and spinal cord.

Microcephalus The congenital smallness of the head.

Neural tube defect A general term for a number of defects which are presumed to have a common origin in failure of the neural tube to develop properly during the embryonic stage. The major conditions include anencephalus, spina bifida, encephalocele.

Omphalocele The membrane-covered protrusion of an abdominal organ through the abdominal wall at the umbilicus.

Patau Syndrome See Trisomy 13.

Reduction deformities of the lower limbs The congenital absence of a portion of the lower limb.

Reduction deformities of the upper limbs The congenital absence of a portion of the upper limb.

Renal agenesis and dysgenesis The failure, or deviation, of embryonic development of the kidney.

Spina bifida The congenital defective closure of the bony encasement of the spinal cord, through which the cord and meninges may or may not protrude. Includes myelomeningocele and meningomyelocele.

Stenosis A narrowing or constriction of the diameter of a bodily passage or orifice.

Tetralogy of Fallot Ventricular septal defect with pulmonary stenosis or atresia, aorta displaced to the right, and hypertrophy of right ventricle.

Tracheoesophageal fistula, esophageal atresia and stenosis An abnormal passage between the esophagus and trachea, or the absence, closure or constriction of the esophagus.

Transposition of the great vessels A congenital malformation in which the aorta arises from the right ventricle and the pulmonary artery from the left ventricle (opposite of normal), so that the venous return from the peripheral circulation is recirculated by the right ventricle, via the aorta, to the systemic circulation without being oxygenated in the lungs.

Trisomy 13 (Patau syndrome) The chromosomal abnormality that is characterized by impaired midline facial development, cleft lip and palate, polydactyly and mental retardation.

Trisomy 18 (Edwards syndrome) The chromosomal abnormality that is characterized by mental retardation, growth retardation, lowset ears, skull malformation and short digits.

Trisomy 21 (Down syndrome) The chromosomal abnormality that is characterized by moderate to severe mental retardation, sloping forehead, small ear canals, flatbridged nose and short fingers and toes.

Appendix B

Number of Deliveries With Selected Birth Defects Which Occurred in the Covered Area to Mothers Not Residing in the Covered Area, 1995

Birth Defect	Total	Region 6	Region 11
Central Nervous System			
Anencephalus	0	0	0
Spina bifida with hydrocephalus	3	3	0
Spina bifida without hydrocephalus	2	1	1
Encephalocele	0	0	0
Microcephalus	2	2	0
Hydrocephalus	5	5	0
Cardiovascular and Respiratory			
Transposition of the great vessels	1	1	0
Tetralogy of Fallot	2	2	0
Hypoplastic left heart	0	0	0
Agenesis or aplasia of lung	1	1	0
Oral Clefts			
Cleft palate alone	3	3	0
Cleft lip with or without cleft palate	3	2	1
Gastrointestinal			
Tracheoesophageal fistula, esophageal atresia/stenosis	0	0	0
Atresia/stenosis of large intestine, rectum or anus	1	1	0
Genitourinary			
Renal agenesis or dysgenesis	2	2	0
Musculoskeletal			
Reduction defects of the upper limbs	1	1	0
Reduction defects of the lower limbs	0	0	0
Diaphragmatic hernia	4	4	0
Omphalocele	1	1	0
Gastroschisis	3	3	0
Chromosomal			
Trisomy 21 (Down syndrome)	5	5	0
Trisomy 13 (Patau syndrome)	0	0	0
Trisomy 18 (Edwards syndrome)	2	2	0
Other			
Fetal alcohol syndrome	0	0	0

Appendix C:

Number of Live Births and Fetal Deaths by Region, Maternal Age, Race/Ethnic Group and Sex

(1) To Residents of PHR 6 and 11, Occurring in 1995

		# LIVE BIRTHS	# FETAL DEATHS
OVERALL (REGIONS 6 & 11 TOGETHER)		111,902	718
BY PUBLIC HEALTH REGION:	6	75,362	516
	11	36,540	202
BY MATERNAL AGE:	<20	18,050	104
	20-24	31,433	169
	25-29	29,438	177
	30-34	21,935	142
	35+	11,036	108
	Unknown	10	18
BY RACE/ETHNIC GROUP:	White	34,088	217
	African American	13,682	152
	Hispanic	60,565	334
	Other / Unknown	3,567	15
BY SEX OF INFANT OR FETUS:	Female	54,790	342
	Male	57,112	371
	Unknown	0	5

(2) Occurrent, Non-Resident Live Births, in 1995

		# LIVE BIRTHS
OVERALL (REGIONS 6 & 11 TOGETHER)		2,503
BY PUBLIC HEALTH REGION:	6	1,491
	11	1,104

Contents

Acknowledgements

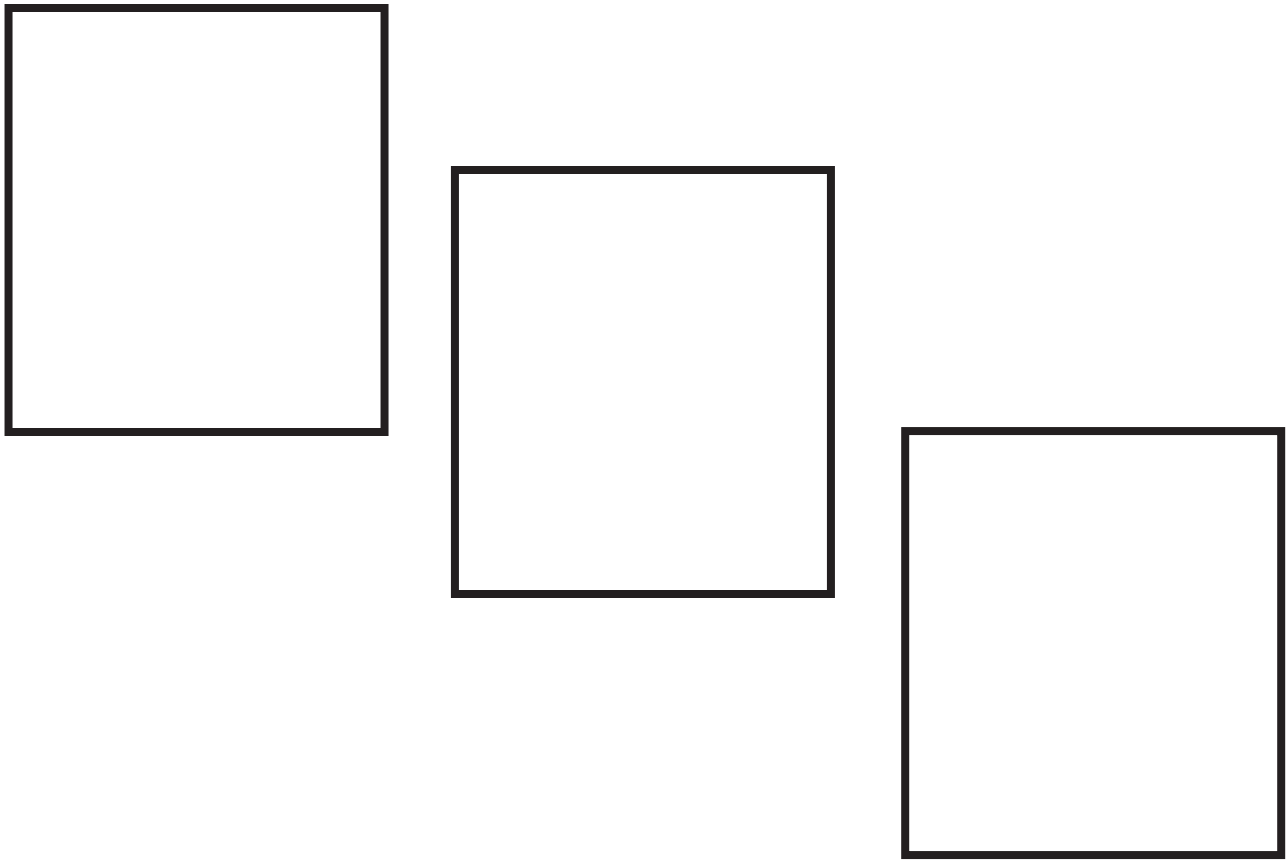
We are grateful for invaluable assistance in setting up the Texas Birth Defects Registry from the Metropolitan Atlanta Congenital Defects Program, Centers for Disease Control and Prevention, and the California Birth Defects Monitoring Program. We also thank the Scientific Advisory Committee on Birth Defects in Texas and our two clinical reviewers, Margaret Drummond-Borg, M.D., and Angela Scheuerle, M.D..

For More Information

This report is a publication of the Texas Birth Defects Monitoring Division, Bureau of Epidemiology, Texas Department of Health, 1100 West 49th Street, Austin TX 78756
(512) 458-7232

Published October 1997

Texas Birth Defects Registry
Report of Birth Defects Among 1995 Deliveries



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